Bypass Surgery for Moyamoya Disease
—Concept and Essence of Surgical Techniques—

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Abstract

This review describes the basic concepts of surgical revascularization for moyamoya disease, including direct and indirect bypass surgery. Direct bypass surgery can improve cerebral hemodynamics and reduce further ischemic events immediately after surgery, but may be technically challenging in some pediatric patients. Indirect bypass surgery is simple and has widely been used. However, its beneficial effects can be achieved 3 to 4 months after surgery, and surgical design is quite important to determine the extent of surgical collateral pathways. Combined bypass procedure, especially superficial temporal artery (STA) to middle cerebral artery anastomosis and indirect bypass, encephalo-duro-myo-arterio-pericranial synangiosis, is a safe and effective option to improve the short- and long-term outcome in patients with moyamoya disease. Alternative techniques are also described for specific cases with profound cerebral ischemia in the anterior cerebral artery or posterior cerebral artery territory. Special techniques to safely complete bypass surgery and avoid perioperative complications are presented, including methods to prevent delayed wound healing, to avoid facial nerve palsy after surgery, and to preserve the STA and middle meningeal artery during skin incision and craniotomy. Finally, the importance of careful management of patients is emphasized to reduce the incidence of perioperative complications, including ischemic stroke and hyperperfusion syndrome.

Key words: moyamoya disease, bypass surgery, surgical technique, direct bypass, indirect bypass

Introduction

Moyamoya disease is an uncommon cerebrovascular disorder that is characterized by progressive occlusion of the supraclinoid internal carotid artery (ICA) and its main branches within the circle of Willis. This occlusion results in the formation of a fine vascular network (moyamoya vessels) at the base of the brain.46 The predominant feature of the pathology of moyamoya disease is now known to be progressive stenosis of the carotid artery terminations, and the moyamoya vessels are the dilated perforating arteries that function as collateral pathways.51 Recent studies have rapidly expanded our knowledge of the basic and clinical aspects of moyamoya disease, including the etiology, pathophysiology, surgical treatment, and long-term prognosis of the disorder.46 In particular, various types of bypass surgery have been developed and are known to improve the long-term outcome in patients with moyamoya disease.3–5,8,13,14,31,33,38,40,44 However, further understanding of the pathophysiology, diagnosis, and treatments of this disease is needed to improve the long-term outcome for these patients.22

In this article, we review recent surgical advances in the treatment of moyamoya disease. Special points of surgical techniques are precisely described. Surgical techniques for specific cases are also presented. Finally, the importance of perioperative management of patients with moyamoya disease is emphasized to reduce the incidence of perioperative complications during and after surgical revascularization.

Superficial Temporal Artery to Middle Cerebral Artery (STA-MCA) Anastomosis and Encephalo-duro-myo-arterio-pericranial Synangiosis (EDMAPS)—Standard, Tips, and Tricks

As we recently reported, STA-MCA anastomosis
and EDMAPS are safe and feasible surgical procedures for pediatric and adult patients with moyamoya disease. These procedures include direct STA-MCA anastomosis and indirect synangiosis. Combined bypass can provide the advantages of both direct and indirect bypass procedures. Clinical results were described previously. Briefly, overall incidences of mortality and morbidity in 123 operations for 75 patients were 0% and 5.7%, respectively. The annual risk of cerebrovascular events during follow-up periods was very low, at 0% in pediatric patients and 0.4% in adults over about 67 months. Here we discuss the theoretical basis and surgical techniques of direct and indirect bypass procedures separately.

**Direct Bypass Procedure**

Direct bypass is useful to improve cerebral hemodynamics and to resolve ischemic attacks immediately after surgery. The frequency of perioperative ischemic stroke is lower after direct or combined bypass than after indirect bypass. Surgery can be technically challenging in a certain subgroup of pediatric patients, because their cortical branches have smaller diameters and are more fragile than those of adults. Therefore, thorough surgical training in vessel anastomosis is essential. To safely complete direct bypass procedures, the following techniques are quite important. First, complete hemostasis of the entire surgical route, including the scalp, muscle, cranium, and dura mater, is essential to prevent disturbance of fine manipulations during the procedures. Elimination of cerebrospinal fluid from the operative field also facilitates direct bypass procedures. Second, clear visualization of the orifice of arteriotomy by staining blue with methylrosaniline chloride (pyoctaninum blue) is quite useful. Blue silicone rubber should be placed beneath the recipient vessel, especially in pediatric patients. Our personal experience suggests that these preparations enable safe STA-MCA anastomosis even in a one-year-old baby (Fig. 1).

Although the incidence of ischemic stroke is lower after direct or combined bypass surgery, recent clinical studies have clarified that careful management of patients is quite important to avoid perioperative complications after bypass surgery because pronounced postoperative changes in cerebral hemodynamics may induce hyperperfusion syndrome, particularly in patients with profound ischemia before surgery. Dramatic postoperative changes in cerebral hemodynamics may also cause rapid diminishment of the basal moyamoya vessels and lead to transient cheiro-oral syndrome or small frontal lobe infarction. Therefore, pre- and postoperative blood flow studies can be important to identify and prevent such serious complications after direct or combined bypass surgery.

Moyamoya disease is often associated with altered cerebral hemodynamics in the frontal lobe, including the territory of the anterior cerebral artery (ACA). However, direct STA-ACA anastomosis is not always essential in all patients with moyamoya disease, probably because the surgical collaterals to the MCA territory may also provide blood flow to the ACA territory through the pial anastomosis. Thus, the collateral blood flow may be redistributed after surgery. However, direct STA-ACA anastomosis is essential in specific patients with pronounced ischemia in the ACA territory, although the number of such cases may be rather small. The frontal branch of STA should be dissected from the scalp to as great a length as possible, which enables easier handling during STA-ACA anastomosis. The frontal branch of STA can be anastomosed to the cortical branch of ACA close to the midline with the usual technique, but the direct bypass procedure should be performed more carefully, because the calibers of both donor and recipient vessels are often smaller than the usual situation in STA-MCA anastomosis (Fig. 2). Postoperative angiography and blood flow studies show improvement or normalization of the cerebral hemodynamics in the involved ACA territory.

The posterior cerebral artery (PCA) is also involved in a certain subgroup of patients with moyamoya disease, and PCA lesions can be observed in approximately 25% to 60% of cases. These patients are considered at higher risk for subsequent ischemic stroke, because the PCA functions as an important collateral route to the ICA territory in moyamoya disease. Some patients are known to develop cerebral infarction in the occipital lobe or temporo-occipital lobe at initial presentation. Surgical revascularization should be planned for both the ICA and PCA territories in these patients. We have developed one-stage bypass surgery that can provide extensive collateral blood flow to the entire hemisphere, as reported elsewhere. Briefly, the technique includes STA-MCA anastomosis targeted to the angular artery and indirect bypass through large craniotomy extended from the frontal to the temporo-parietal area. Follow-up cerebral angiography reveals that surgical collaterals supplied blood flow widely to the operated hemispheres including the posterior temporal and parietal lobes. Postoperative blood flow studies also demonstrated marked improvement of cerebral hemodynamics.

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Delayed wound healing or scalp necrosis is known as one of the most serious complications after STA-MCA anastomosis in patients with moyamoya disease.\textsuperscript{19,29} We have used a modified surgical technique to dissect the STA to avoid this problem for 15 years. No serious complications of wound healing and scalp necrosis have occurred. First, the STA branches are carefully dissected from the surrounding galeal tissue under the surgical microscope. The dissected STA should be “naked,” because the surrounding galeal tissue is quite important for wound healing. Next, the galeal “track” is always repaired after STA dissection by suturing the galeal tissue. The small amounts of time and effort contribute to the preservation of scalp blood flow, thus supporting wound healing (Fig. 4). The technique can be applied to STA-MCA anastomosis for patients with atherosclerotic carotid artery diseases (Kuroda et al., unpublished data).

In patients with advanced-stage moyamoya disease, the vault moyamoya vessels are frequently identified through the middle meningeal artery (MMA) and STA.\textsuperscript{21} Therefore, the STA branches should be preserved during surgical revascularization if these vessels are involved in the vault moyamoya vessels. A representative case is presented in Fig. 5. This 52-year-old male had spontaneous collaterals to the ACA branches through the frontal branch of STA. Thus, the frontal branch of STA was preserved and used as the donor artery for encephalo-arterio-synangiosis by positioning on the brain surface.

As described above, direct STA-MCA and/or STA-ACA anastomoses are considered to contribute to
Fig. 3   Radiological and intraoperative findings of a 38-year-old female who developed ischemic stroke.  
A: Diffusion-weighted magnetic resonance (MR) image demonstrating cerebral infarction in the left posterior temporal and occipital lobes at onset. 
B: MR angiogram revealing severe stenosis of the left posterior cerebral artery (arrow) as well as occlusive lesions in the bilateral internal carotid arteries. 
C: Preoperative oxygen-15 gas positron emission tomography (15O-gas PET) scans showing marked reduction of cerebral blood flow in the entire left hemisphere (left column). Oxygen extraction is elevated in the left parieto-occipital lobe (right column, arrowhead). 
D: Intraoperative photograph showing a wide craniotomy exposing the posterior temporal and parietal lobes. The parietal branch of the superficial temporal artery (STA) is anastomosed to the angular artery to directly supply blood flow to the oxygen extraction-elevated area (arrow). The frontal branch of STA is anastomosed to the prefrontal artery (arrowhead). 
E: Postoperative three-dimensional skull computed tomography scan demonstrating the extent of craniotomy. 
F: Postoperative 15O-gas PET scans showing improvement of the cerebral hemodynamics (left column) and oxygen metabolism (right column) in the operated hemispheres.

Fig. 4   Intraoperative photographs of left superficial temporal artery (STA) to middle cerebral artery double anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis for a 6-year-old boy who developed transient weakness of the left extremities. 
A: The main trunk of STA and its two main branches are carefully dissected from the surrounding galeal tissue under surgical microscope. Note that there are no galeal tissues around the dissected STA branches. 
B: The main trunk of STA and its frontal and parietal branches are still patent after complete dissection. Note that dissection of the STA branch resulted in the track-like galeal injury, where subcutaneous fatty tissue is exposed (arrow). 
C: After STA dissection, the galeal injury should be carefully repaired to preserve blood flow in the scalp, using absorbable surgical sutures (arrow). 
D: The galeal injury after dissection of the frontal STA branch is completely repaired, contributing to good wound healing (arrows).

A good short- and long-term outcomes of patients with moyamoya disease. However, the course of the frontal STA branch should carefully be checked prior to surgery. As reported before, the frontal branch of STA runs tortuously upward and forward to the forehead, where it supplies the muscles, integument, and pericranium in this region, and anastomoses with the supraorbital and frontal arteries. In a certain subgroup of patients, it runs in an extremely caudal direction. In such cases, full dissection of the frontal STA branch may injure the temporal branch of the facial nerve and cause postoperative palsy of the frontalis muscle. Therefore, only the distal portion of the frontal STA branch should be dissected from the scalp to avoid postoperative frontalis palsy, if the course is extremely caudal. The artery close to the temporal branch of the facial nerve should be left intact (Fig. 6). The dissected length is enough for direct bypass to the frontal branches of the MCA. After STA-MCA anastomosis, the frontal branch of STA can be guided into the intracranial space through the burr hole at the pterion (Fig. 6).

Indirect Bypass Procedure

Surgical procedures for indirect bypass are specific for moyamoya disease. Indirect bypass surgery that induces spontaneous angiogenesis between the brain surface and the vascularized donor tissues is technically simple to do and has been widely used. Donor tissues include the STA, dura mater, temporal muscle, and galeal tissue. However, several important issues should be considered when performing indirect bypass procedures. First, the beneficial effects are not immediate because surgical collaterals require 3 to 4 months to develop.
Fig. 5 Intraoperative photographs of superficial temporal artery to middle cerebral artery (STA-MCA) single anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis for a 52-year-old male who developed minor ischemic stroke. A: Preoperative right external carotid angiogram showing the collateral circulation to the anterior cerebral artery branches spontaneously developed through the frontal branch of STA (arrows). B: Skin incision (black line) and course of two branches of STA (red lines). Note that the frontal branch of STA (arrow) is crossing the line of the skin incision. C: The STA branches are carefully dissected, and the scalp flap is reflected. Then, the temporal muscle and frontal pericranial flap are dissected. The frontal branch of STA is still intact (arrow). D: The dura mater is opened, leaving the middle meningeal artery branches intact. The parietal branch of STA is anastomosed to the cortical branch of MCA (arrowhead). The frontal branch of STA is still intact (arrow). E: The dural window is covered by the temporal muscle (M) and frontal pericranium (P). Note that the frontal branch of STA is still intact (arrow) and is used as the donor artery for encephalo-arterio-synangiosis. F: Postoperative right external carotid angiogram demonstrating collateral blood flow through the STA-MCA anastomosis and indirect bypass. Note the preserved frontal branch of the STA (arrows).

suggesting that there is a potential risk of perioperative ischemic stroke.\textsuperscript{9,41} Second, previous studies have demonstrated that collateral pathways through indirect bypass extensively develop in almost all pediatric patients, but not in about 40% to 50% of adult patients.\textsuperscript{36} In fact, combined, but not indirect, bypass surgery could reduce the incidence of re-bleeding in adult moyamoya disease.\textsuperscript{15} Third, surgical design is quite important because the extent of surgical collateral pathways depends on the size of the craniotomy and the extent of the indirect bypass. Thus, the revascularized area is confined to the craniotomy field after indirect bypass. Recent multivariate analysis has proven that “small craniotomy” surgery can be an independent predictor for poor intellectual outcome in pediatric moyamoya disease, probably because of persistent cerebral ischemia in the frontal lobes even after surgery.\textsuperscript{23} Figure 7 shows representative cerebral angiography and three-dimensional skull computed tomography findings after various types of indirect bypass surgery, demonstrating that indirect bypass through a smaller craniotomy develops less extensive surgical collaterals.

Based on these observations, we have recently developed a novel indirect bypass procedure by using the vascularized frontal pericranial flap, named EDMAPS (see above).\textsuperscript{24} The frontal pericranial flap is large enough to widely cover the frontal lobe (Figs. 2, 3, and 5). Postoperative cerebral angiography and blood flow studies have shown that the pericranial flap functions well as a donor tissue for indirect bypass, especially in pediatric patients with
Fig. 7 Postoperative external carotid angiograms (A–C) and three-dimensional skull computed tomography scans (D–F) showing the relationship between craniotomy size and the extent of surgical collaterals through indirect bypass after encephalo-duro-arterio-synangiosis (A, D), encephalo-myo-synangiosis (B, E), and encephalo-duro-arterio-myo-synangiosis (C, F).

moyamoya disease. Figure 8 demonstrates the representative radiological findings before and after STA-MCA single anastomosis and EDMAPS in an adult patient with hemorrhagic onset. Surgical collateral pathways have extensively developed and provide blood flow widely to the operated hemisphere. Finally, the basal moyamoya vessels are markedly diminished.

The MMA can function as one of the important surgical collaterals through the dura mater. Therefore, the MMA should carefully be preserved during craniotomy. However, the course of the anterior (frontal) branch of the MMA in the region of lesser wing of the sphenoid and adjacent parietal bone greatly varies in adults. Generally, the anterior branch of the MMA is believed to run within the groove in the medial surface of bone, but this pattern is observed in less than 30% of adults. Alternatively, the course of the anterior branch of the MMA is completely enclosed within a bony canal in the lesser wing of the sphenoid and parietal bone in about 50% to 75% of adults. Therefore, the MMA can easily be damaged during usual fronto-temporal craniotomy. As shown in Fig. 8G and H, we have modified the design of fronto-temporal craniotomy to avoid this problem. The MMA can be kept intact by carefully drilling out the bone surrounding the MMA to avoid damage. Figure 8E and F demonstrate that the dilated anterior branch of the MMA remains intact and functions as one of collateral pathways even after surgery.

Fig. 8 Radiological findings of a 55-year-old female who developed transient ischemic attacks followed by right thalamic hemorrhage. She underwent right superficial temporal artery to middle cerebral artery (STA-MCA) single anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis (EDMAPS) safely. A: Postoperative three-dimensional skull computed tomography scan showing the extent of craniotomy. B, C: Pre- (B) and postoperative (C) right internal carotid angiograms revealing marked diminishment of the basal moyamoya vessels after surgery (C, arrows). D–F: Pre- (D) and postoperative (E, F) right external carotid angiograms showing extensive developments of the surgical collaterals through the STA-MCA anastomosis (E, F; arrow) and indirect bypass. Note that the middle meningeal artery (MMA) is preserved even after surgery (D–F, arrowhead) and that the deep temporal artery has increased diameter after surgery (E, asterisk). G, H: Intraoperative photographs of right STA-MCA single anastomosis and EDMAPS. Fronto-temporal craniotomy is designed to avoid injury of the MMA during craniotomy (G). The MMA can be preserved intact by carefully drilling out the bone surrounding the MMA (H).

Conclusions

In this article, we describe the basic concepts of surgical revascularization for moyamoya disease. In particular, STA-MCA anastomosis combined with a novel indirect bypass, EDMAPS, can be a safe and effective procedure for pediatric and adult patients with moyamoya disease. However, it is quite im-
important to achieve the optimum effects by modifying the surgical procedures according to the cerebral hemodynamics and spontaneous collateral pathways in each case. Also, it is essential to understand the anatomy of the scalp, STA, and MMA to avoid surgical complications and improve outcome. In addition to surgical techniques, careful management of the patients is critical to reduce the incidence of perioperative complications, including ischemic stroke and hyperperfusion syndrome.

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References


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